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Review Article

Dextrocardia with Situs Solitus

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Abstract

Congenital cardiac diseases are responsible for about 1% of the whole of this organ's illness. Until recently, they were conditions related to pediatric care, since few patients grew up to adulthood. The human aging added to improvement of recognition and treatment on childhood shifts it towards a number of adults with congenital pathologies. It is necessary to the adult cardiologist being prepared to recognize congenital conditions and care of population with these conditions associated to aging diseases. This phenomenon is challenging and demands to the cardiologist new knowledgements. In this article, it is reviewed a rare situation: dextrocardia with situs solitus.

Keywords: dextrocardia; dextroversion; situs solitus; congenital cardiac disease

Introduction

Pediatric cardiology is a recent issue and it had been born because of a lot of knowledgement acquired over the years that had to be refined after the advent of cardiac surgery [1]. Previously, the mortality rate of this condition was high with few of child getting adulthood. Nevertheless, the progression of surgical and percutaneous treatments contributed with increase of survivorship of this population [2].

The overall prevalence of cardiac heart defects is approximately 1% [1] and it may be rarer when it is evaluated only severe conditions. Aging rises the risk of diseases related with elderly, as hypertension, coronary diseases and stroke [3]. The association of congenital and adquired diseases may make it difficult to the physicians to manage them.

A lot of information has been recently produced and show the prevalence of congenital heart disease may be increasing, probably related to greater use of diagnostic methods, as echocardiography. Single conditions, as atrial septal defects, in opposite of complex disease, are responsible to the elevation [4].

Otherwise, the number of adults with congenital heart disease are is growing, and it is related to effectiveness of clinical care and diagnosis [5], reforcing the value of undestanding these conditions.

In this article, we review isolated dextrocardia with situs solitus.

Definitions:

Dextrocardia (or dextroversion) is a cardiac malposition defined as a location of the heart in the right side of the thorax, with the apex directed to the right side. This condition must be differentiated with dextroposition, when the malposition occurs because of extra-cardiac problems, as athelectasia, tumors, left-sided pleural effusion or pneumothorax [6]. The incidence of all types of dextrocardia is about 0,8/10000 pregnancies [7].

This malaposition of the heart may occur with situs solitus, situs inversus or situs ambiguus. When it occurs with no other alteration, it Is denominated isolated dextrocardia. Another classification divides into complicated, when it's related to a disturbance of heart circulation, and non-complicated, when no abnormalities of valves, septum, great vessels and circulation are founded [8].

"Situs" is a term used to assign the spacial relation of atria and visceral organs. When the right atria are located in the left side, it is called atrial situs inversus. It may happen with visceral organs too, as example the liver at the left side and stomach and spleen at the right side – abdominal situs inversus [1,9]. We called situs inversus totalis the combination of atrial plus abdmonial situs [1,9,10].

The determination of atrial situs should be considered analyzing systemic venous conection (inferior venous cava draining to right atria) and morphology of atrial appendage. Right atria appendage is triangular and has a thick wall [9].

Clinical Presentations:

The clinical diagnosis of dextrocardia is made with identification of right-side apex and shifted cardiac and murmur position [8,9].

When dextrocardia is associated with atrial situs solitus and normal atrioventricular concordance, it means true dextrocardia. There is no anatomic alteration beside rotation of the heart to the right. The atrioventricular relationship is normal [9].

This condition may be related to ventricular septal defects, aortic coarctation and atrial septal defects [7,9,10,11]. In a cohort evaluating twelve thousand pregnancies, atrial septal defects, ventricular septal defects, double-outlet left and right ventricle and abnormal pulmonary valve were the most

common cardiac associations and lung and urogenital abnormalities the most common extra-cardiac one [7]. A structurally normal heart is very rare, with incidence of 1:900000 cases in adults [10]. The clinical presentation depends on the presence of association with another anatomic alterations per six.

Dextrocardia associated with ventriculoarterial discordance is compatible with transposition of Great arteries. It may occur as well when dextrocardia is associated with atrioventricular and ventriculoarterial discordance and atrial situs solitus - Congenitally Corrected Transposition of Great arteries. This condition is the most common malformation related with dextrocardia and atrial situs solitus [1]. There may be mesocardia. Other cardiac malformations, as ventricular septal defects, are common. It may be occurred double inlet left ventricular with hypoplasic right ventricle and other malformations [11].

Complementary analysis:

A chest radiography is enough to identify dextroversion (Figure 2).

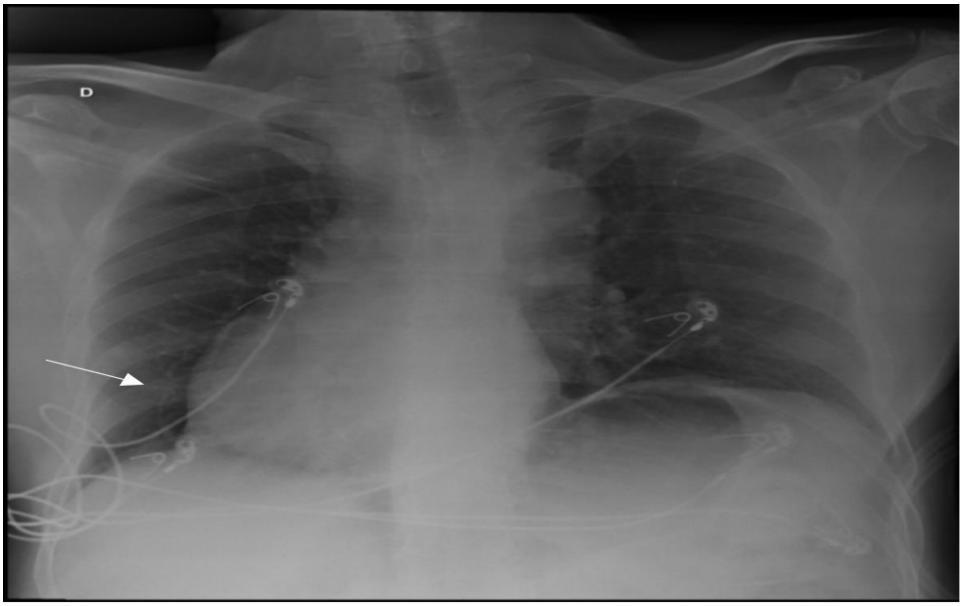

Figure 2: A chest radiography shows apex turned to the right.

It is important to search pulmonary abnormalities may turn the heart to the right, as pleural effusion or hernia, for example. An electrocardiographic analysis of these conditions shows normal P-axis and right QRS axis (the shortening of QRS amplitude from V1 to V6) (Figure 3).

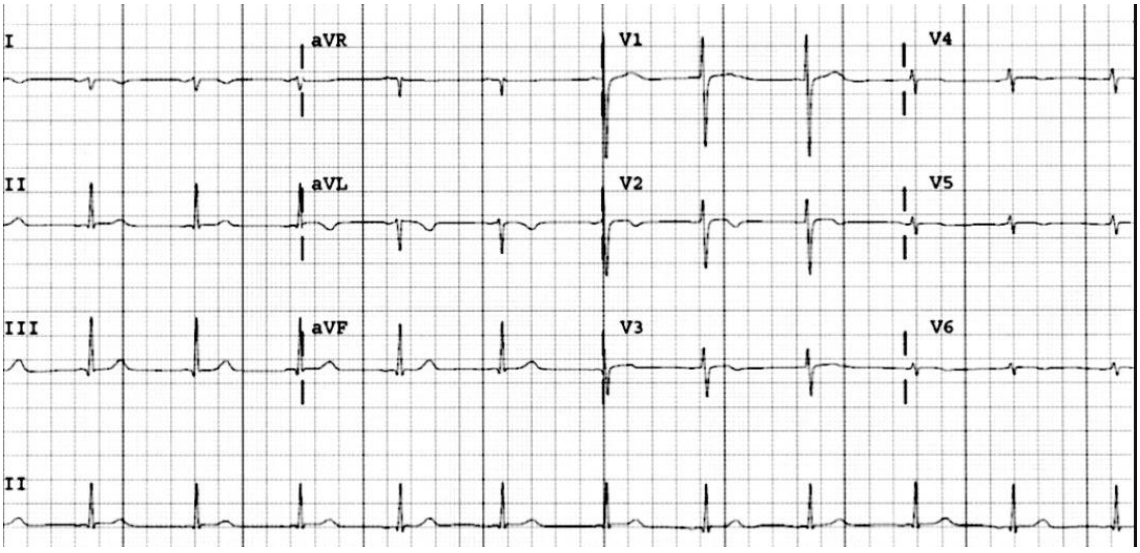

Figure 3: An electrocardigram in dextrocardia. The axis is turned to the right (leads I is negative and avF is positive). P wave is negative at leads I. QRS is shortening from V1 to V6.

Echocardiography, Cardiac tomography (Figure 4) and cardiac magnetic resonance are the best methods to evaluate the heart, including morphology, atrial and visceral situs, atrioventricular concordance, ventriculoarterial concordance, valve morphology and functioning. Echocardiography is the most common method made and must be done in all suspected patients.

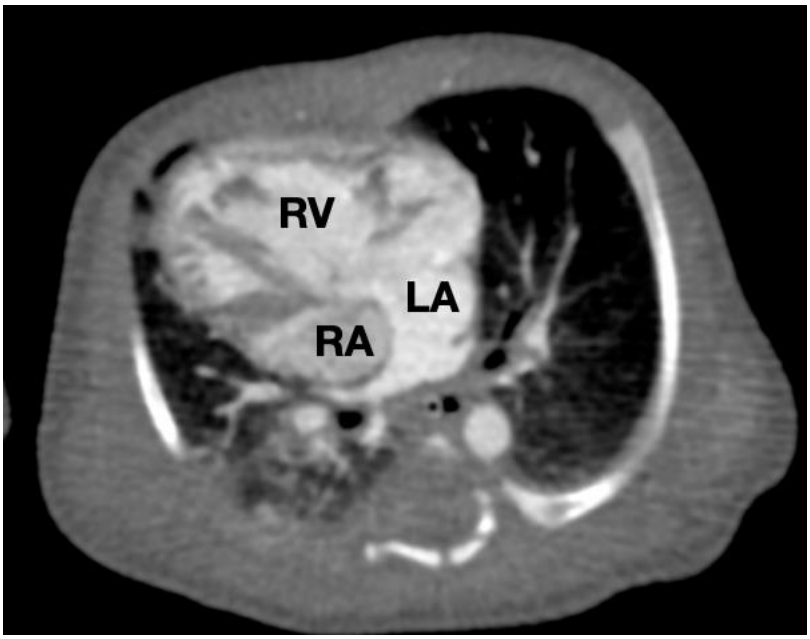

Figure 4: Complex congenital cardiac disease is shown. A case of dextrocardia with situs solitus in a patient with atriovcentricular discordance and mitral atresia. RV: right ventricle, LA: left atria, RA: right atria.

Isolated dextrocardia and situs solitus:

This condition is very rare, as cited above (1:90000 cases), and may be identified at the adulthood [12]. In this case, the growth may be normal, and diagnosis made occasionally. It's important to identify adults with others cardiovascular diseases, as dyslipidemia, hypertension, diabetes melitus and coronary artery disease.

We previously described a case of an adult patients admitted with acute coronary syndrome in a patient with dextrocardia and situs solitus (no other cardiac abnormalities) successfully treated with percutaneous coronary intervention [11]. It is interesting to describe Goel technique, that consists of inverting cine images (mirror image) during coronary catheterization, making easier to do the exam [12]. About cardiac catheterization in patients with dextrocardia, there are several hints to overcome the procedure: image acquisition wit mirror image (left to right anterior oblique view), reversal digital technique (Goel), using transfemoral access may get easier catheterization, using non-common catheters [13].

There are many publications related to interventions in patients with dextrocardia with situs solitus, as percutaneous closure of atrial defects [13].

Conclusion:

Dextrocardia is a rare malposition of the heart and may be associated with misplace of the atria (situs inversus) and others malformations. Considering the growing survivorship of congenital cardiac disease, adult`s cardiologists must be familiar to some types of congenital diseases. Isolated dextrocardia is a very rare phenomenon.

Acknowledgments:

None.

Conflicts of Interest:

No conflict of interest.

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